Outcomes Evaluation in Pulmonary Rehabilitation for Patients with Cystic Fibrosis at Nationwide Children's Hospital

UNDERGRADUATE RESEARCH THESIS

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Abstract

INTRODUCTION: Pulmonary rehabilitation (PR) is an individualized program aimed at helping patients manage and cope with their symptoms. Comprehensive PR includes education regarding pulmonary disorders, exercise training, nutrition classes, and psychosocial counseling. Traditional PR programs for patients with chronic respiratory diseases such as COPD have shown positive outcomes. In addition, physical activity has been shown to benefit cystic fibrosis (CF) patients. As a result, PR may be a beneficial intervention for CF. However, there are few centers that have designated programs for CF patients and there are no current studies describing the outcomes of pulmonary rehabilitation programs for CF patients. The purpose of this research was to conduct an outcomes evaluation of an AACVPR-accredited PR program for CF patients. METHODS: The study examined clinical and psychosocial data of all CF patients that have participated in the program including pre and post 6-minute walk tests (6MW), pulmonary function tests (PFT), body mass index (BMI), and health-related quality of life (HRQOL). It also examined if differences exist based on age, severity of disease, program compliance, and sex. There was found to be a significant difference in pre/post 6MW results ($p = .001$), and a significant difference in pre/post BMI values ($p = .002$). There was found to be a significant difference between pre and post 6MW distances for the varying levels of disease severity ($F(5,27) = 2.838$, $p = 0.035$), suggesting pulmonary rehabilitation may be more beneficial for CF patients with mild, moderate, or moderately-

severe lung impairment. Female patients were also shown to have a greater change in BMI from pre to post program than were male patients ($F(1,34) = 4.160$, $p = 0.049$). There was no significant change in the FEV1 and FEV1/FVC PFT values from pre to post program. Tests on other variables were not performed due to incomplete records. CONCLUSIONS: Improvement in analyzed variables from pre to post PR, indicate that CF patients likely benefit from PR much like others with chronic respiratory diseases.

Dedication

This document is dedicated to my family.

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M.M. Monasky, D.M. Taglieri, **A.K. Jacobson**, K.M. Haizlip, R.J. Solaro, P. Janssen. Post-Translational Modifications of Myofilament Proteins Involved in Length-Dependent Prolongation of Relaxation in Rabbit Right Ventricular Myocardium. *Biophysical Journal;* 102(3) pp. 614a.
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Fields of Study

Major Field: Respiratory Therapy

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Chapter 1: Introduction and Statement of the Problem

Cystic fibrosis (CF) is a genetically inherited disease that causes the production of abnormally thick and sticky mucus which builds up in the lungs and the pancreas. This buildup of mucus can lead to respiratory infections and digestion problems. CF affects over 40,000 people in the United States and 70,000 people worldwide (Cystic Fibrosis Foundation, 2010). In the past, people with CF rarely lived to attend elementary school; however over the past three decades, due to medical advancements, the average life expectancy for CF has increased to 37 years (Zieve & Hadjiliadis, 2011). Although CF patients are living longer, they still face many chronic symptoms that affect their health status and quality of life. Some symptoms include increased mucus production, recurrent pulmonary infections, inability to gain weight normally throughout childhood, fatigue, and increased sinus pressure. There are many medical treatments available to CF patients that have been helpful in treating some of these symptoms.

Mathews, et al emphasize that because cystic fibrosis is a genetically inherited disease, it is a lifelong condition. The primary process of the condition that affects the lung is not the constitution of the mucus produced in the lungs; rather, it is the inability of the lungs to effectively clear the secretions. The buildup of secretions in the lungs causes obstruction which impairs overall respiratory function. The accumulation of secretions in the lungs invites bacterial growth which can cause increased impairment and inhibition of mucociliary clearance. Bacterial growth causes chronic respiratory infections in CF

patients. To treat and prevent mucus accumulation in the lungs chest physiotherapies are utilized to mobilize secretions and improve secretion clearance. Physical activity has been important in prophylactically mobilizing secretions in day to day life as well as in the hospital settings (Mathews et al, 1964). Physical activity and training has been found to help improve quality of life, and may slow the deterioration of lung function in pediatric patients as well (Bradley & Moran, 2008).

> Pulmonary rehabilitation has been defined as:
> "an evidence-based, multidisciplinary, and comprehensive intervention for patients with chronic respiratory diseases who are symptomatic and often have decreased daily life activities. Integrated into the individualized treatment of the patient, pulmonary rehabilitation is designed to reduce symptoms, optimize functional status, increase participation, and reduce health care costs through stabilizing or reversing systemic manifestations of the disease"(Nici et al, 2006).

According to Sharma, et al pulmonary rehabilitation helps to improve patients' quality of life and maximize their capabilities. Pulmonary rehabilitation is a tailored program aimed at helping patients cope and manage their symptoms (Sharma et al, 2010). The program can include education regarding pulmonary disorders, exercise training, nutrition classes, and psychosocial counseling. Research studies dating back to the mid-1970s have shown that pulmonary rehabilitation programs have had success increasing exercise tolerance and quality of life in COPD patients by incorporating both exercise, education, and breathing retraining (Troosters et al, 2005). Because of the known successes of traditional pulmonary rehabilitation programs geared toward patients with chronic respiratory diseases, such as COPD, and the known benefit of physical activity in CF patients, pulmonary rehabilitation is also a treatment for CF patients. Though pulmonary rehabilitations programs might be beneficial for CF patients, there are few

centers that have a designated program for CF patients and there are no current studies describing the outcomes of pulmonary rehabilitation programs for CF patients.

The purpose of this study was to perform an outcomes evaluation of a pulmonary rehabilitation program for cystic fibrosis patients at Nationwide Children's Hospital in Columbus, Ohio. The study examined clinical and psychosocial data of all CF patients that have participated in the program including pre and post 6-minute walk tests (6MW), pulmonary function test (PFT) results, body mass index (BMI), and health related quality of life (HRQOL). This study also examined if differences exist based on age, severity of disease, program compliance, and sex.

Chapter 2: Related Research

A literature search was conducted in Medline/ Pubmed and Academic Onefile databases using the keywords pulmonary rehabilitation, cystic fibrosis, and exercise. The following is a review of current literature related to the proposed research. In 2008, Bradley & Morgan conducted a review to determine if a prescribed exercise routine would help improve CF patients' lung function and slow the deterioration of clinical and physiological symptoms. The researchers reviewed all available literature, which totaled seven different studies, regarding physical exercise and its effects on CF patients. They found there is evidence that both aerobic and anaerobic physical training has positive effects on exercise capacity and lung function. This is the evidence of both short term and long term studies that apply to the general CF patient population. The review also provided some evidence that the beneficial effects from physical activity persist for a period of time even after the activity is stopped; however, the exact period of time is unknown. The review ultimately supports that physical activity and training has been found to help improve quality of life and may slow the worsening of lung function in CF patients; additionally there is no evidence that physical activity and training should not be included in the care plan of CF patients (Bradley & Moran, 2008).

Orenstein, et al conducted a study to determine the "effects of home-based, semi-supervised, upper body strength training regimen with a similarly structured aerobic

training regimen" in CF patients. Sixty seven patients ranging in age from 8-18 years were randomized into either an anaerobic training regimen using an arm ergometer with weight resistance or an aerobic training regimen with a stair stepper. Patients were told to exercise three times a week for one year. Measurements of pulmonary function, quality of life, strength, and aerobic fitness were recorded at baseline, 6 months, and 12 months. Neither aerobic nor anaerobic exercise proved to more beneficial than the other. However, there was a significant improvement in upper and lower body muscle strength for both aerobic and anaerobic exercise after 6 and 12 months. Additionally, patients engaging in both aerobic and anaerobic exercise over 6 and 12 months had a significant weight gain (Orenstein et al, 2004).

A study performed by Nixon, et al aimed to study the prognostic value of exercise testing in CF patients. The investigated revealed a strong correlation between survival of patients and aerobic fitness measured by V02 max. The study also showed that after aerobic training patients have greater cardiovascular efficiency and ventilatory muscle strength. The study also factored in the growth of *P. cepacia* in the lungs, which was found to be an independent cause of mortality in CF patients. In CF patients that were not colonized with *P. cepacia*, lack of aerobic fitness was found to be the only significant independent cause of mortality. The results support that exercise testing should be valued as a tool to evaluate CF patient prognosis; aerobic fitness was strongly correlated with 8 year survival rates (Nixon et al 1992).

Moeller, et al performed a study to examine the effects of a pulmonary rehabilitation program with components of exercise and chest physiotherapy on airway

inflammation in CF patients. Eighteen patients with stable CF were obtained and enrolled in a three week inpatient rehabilitation program in Davos, Switzerland. The patients' symptoms were assessed at the beginning and end of the three weeks and were used to determine a clinical score based on the cystic fibrosis clinical score. Patients were also assessed at baseline and post program by pulmonary function testing, exhaled breath condensate, and sputum analysis. Throughout the three weeks the patients had twice daily inhaled treatments as well as chest physiotherapy administered by a healthcare professional. They participated in an exercise program lead by a pediatric sports therapist. The physical exam after the three weeks compared to the beginning of the three weeks resulted in an improved clinical score, as well as improved inspiratory capacities and forced vital capacities. The study found no significant difference in the number of sputum cells or the sputum cytokines or markers within the three weeks. The study ultimately provided evidence that a short term rehabilitation program for CF patients significantly improves subjective clinical symptoms as well as lung function in the FVC and IC, but has no effect on airway inflammation or obstruction Moeller et al, 2010).

A majority of the research about the effectiveness of pulmonary rehabilitation has been focused on the COPD patient population. A 2004 study performed by the California Pulmonary Rehabilitation Collaborative Group sought to determine the outcomes of pulmonary rehabilitation programs in California in reducing dyspnea, improving quality of life, and reducing the utilization of healthcare resources by compiling patient assessment data from nine centers throughout California. Of the patients that were

included in the study 93% of 522 total patients were diagnosed with COPD. The study showed significant improvement in reduction of dyspnea and quality of life measures through a mean change in the Shortness of Breath Questionnaire measurements and the Physical Component Scores of the Medical Outcomes Short Survey Form. The study also showed over 18 months of follow up that pulmonary rehabilitation programs led to a significant reduction in healthcare utilization including decreased hospitalization days, urgent care visits, physician office visits, and telephone calls to physicians (California Pulmonary Rehabilitation Collaborative Group, 2004).

Reis, et al performed a study in 1995 to compare the physiological and psychosocial effects of providing solely education verses a comprehensive pulmonary rehabilitation program to patients with COPD. The study took place at a university hospital with 119 stable COPD patients. The patients were randomly assigned to either an 8 week education program or an 8 week pulmonary rehabilitation program; all patients were then tracked for 6 years. Patients that were enrolled in the pulmonary rehabilitation program verses the education program has significant improvements in maximal exercise tolerance, maximal oxygen intake, breathlessness, muscle fatigue, and self-efficacy for walking. There was no significant difference between the pulmonary rehabilitation patients and the education patients in lung function, quality of life, hospital days, or depression. The patients enrolled in the pulmonary rehabilitation program did have an increased survival rate; however, it was not a significant difference from the patients that received purely education. The results of the study strongly support the benefits obtained from pulmonary rehabilitation in the COPD patient population (Reis et al. 1995).

Griese et al. performed a retrospective chart review of rehabilitation data of cystic fibrosis patients in Israel, Germany, and Switzerland of 142 patients and 240 different rehabilitation stays. The patients from Germany and Switzerland were combined in one group to compare to the patients in Israel who underwent rehabilitation in a different climate. The study found that in both groups, patients' weight and lung function did significantly improve in CF patients of all ages. The rehabilitation sites in Israel found improvements in FVC and oxygen saturation, whereas the rehabilitation sites in Germany found improved FEV1 measurements. However, this review was unable to compile various pieces of data and complete records because many of the pulmonary rehabilitation programs failed to collect and report all important pre and post patient data related to patient outcomes in the program. The review does highlight the importance and need for pulmonary rehabilitation programs to track and maintain detailed records for necessary outcome analyses (Griese et al., 2010).

Chapter 3: Methodology

Study and Research Objectives

The purpose of this study was to examine outcomes of the pulmonary rehabilitation program for patients with cystic fibrosis at Nationwide Children's Hospital in Columbus, Ohio. The following questions were addressed in this study:

1. Does a comprehensive pulmonary rehabilitation program for adult and pediatric patients with cystic fibrosis change outcomes including:
    a. health-related quality of life
    b. 6-minute walk distance
    c. body mass index
    d. pulmonary function
2. Do outcomes for CF patients differ based on:
    a. age
    b. disease severity
    c. program compliance
    d. sex

The Cardiopulmonary Rehabilitation Program at Nationwide Children's

The Cardiopulmonary rehabilitation program at Nationwide Children's began in 2005 and became the first pediatric American Association of Cardiovascular and

Pulmonary Rehabilitation (AACVPR) certified pulmonary rehabilitation program in 2009. Conditions that qualify for the program include obstructive lung disease such as CF, asthma; restrictive pulmonary disease; other pulmonary conditions, such as pre/post-transplant, pulmonary hypertension, sickle cell disease; and cardiac conditions/diseases. However, this study focuses particularly on CF patients that participated in the program. A typical program lasts 6-8 weeks, meeting 2-3 times each week, with each session lasting 1.5-2 hours; however, the program can be extended on an individual basis, and some chronic patients can re-enrolled if need be.

The program consists of a multidisciplinary team comprising physical therapy, therapeutic recreation/occupational therapy, massage therapy, nutrition, psychology, and education. Physical therapy meets with the patients each visit working to improve cardiovascular endurance, increase chest mobility and improve breathing techniques, and to improve strength through upper and lower body strength training. Therapeutic recreation with meet with a patient 2 times a week if the patient attends 3 sessions a week, or once a week if they attend 2 sessions a week. Therapeutic recreation works with patients to help teach them to teach time management with health considerations, to help develop healthy leisure activities, to gain community resources, and to help manage stresses involved with participating in leisure activities. Massage therapy meets with patients once a week to provide pain relief, relaxation, and increased circulation. Nutrition meets once a week with patients to analyze food recall for calories, fat, protein, vitamins and minerals in diet. They provide individualized patient education and nutrition goals and will recommend supports for lifestyle and dietary changes.

Psychology meets once a week with patients to teach relaxation techniques to cope with symptoms and generally work to improve quality of life and mental status. For CF patients, education is provided by a register respiratory therapist and can be given at any point during their program at least once. Each of the areas of the Cardiopulmonary Rehabilitation program is tailored for each specific patient.

In order to participate in the program at Nationwide Children's patients need to be referred by an MD based on qualifying criteria. Patients with certain disease/ conditions can be referred as listed above; patients can also be referred based on symptoms. Examples of physical symptoms include decrease in physical activity; decreased functional status; increased shortness of breath, dyspnea, and fatigue; decrease in pulmonary function; nutrition exhaustion or obesity. Psychosocial symptoms could be reduced health related quality of life or disease effect on psychosocial status. To additionally qualify for the program patients have to be greater than 4 years of age and able to communicate and follow directions. Patients must also have reliable transportation and be willing to travel to the hospital's main campus.

The population in this study was the 41 total CF patients that have completed the pulmonary rehabilitation program to date at Nationwide Children's Hospital in Columbus, OH. This population includes both pediatric and adult patients.

Study Design and Procedure

A retrospective chart review was performed of all CF patients that completed the Pulmonary Rehabilitation program at Nationwide Children's Hospital dating back to 2005. Patient data was reviewed at the patients' baseline and then at the completion of

their pulmonary rehabilitation program. The study compared pre and post program clinical and psychosocial data of CF patients in the program including physical training measured by pre and post 6MW distance, PFT results, BMI, health-related quality of life measured by responses on standardized instruments, and overall compliance with the program. This study also examined if differences exist by age, severity of disease as measured by percent predicted FEV1, and compliance of the patients that participated in the program. Program compliance was defined as completion of the pulmonary rehabilitation program.

Statistical Analysis and Instrumentation

A database was created using SPSS® (Statistical Package for the Social Sciences) Version 19 containing the data collected via the survey instrument. Dummy-coding was used to record nominal and ordinal data in an effort to facilitate statistical analysis. Data was compiled from the medical records of CF patients pre-program and post-program. Health related quality of life (HRQOL) scores were evaluated through survey responses on standardized instruments. The HRQOL survey instrumentation utilized by the program is the SF-36. 6MW tests are measurements of exercise capacity. The test is measured by the total distance in feet a patient can walk in six minutes. A pre and post weight and height was collected for all patients; this was then used to calculate a pre and post BMI (kg/m^2). In order to determine the severity of the patients' disease states pre and post data and PFT results were reviewed. Pre and post PFT results were measured as close to the start and completion date of the program as possible; all collected PFT data were within three months before the start of the program and within three months after

completion. The FEV1 % predicted values of the pre data were used to determine each patient's severity of disease based on their obstruction. Using Wilkin's *Egan's Fundamentals of Respiratory Care* patient were classified as either normal, mild, moderate, moderately-severe, severe, or very severe as demonstrated in the table below (Wilkins, 2009).

Degree of Impairment	FEV1 % predicted
Normal	80%-120%
Mild	70%-79%
Moderate	60%-69%
Moderately-Severe	50%-59%
Severe	35%-49%
Very Severe	<35%

Table 1: Severity of Disease Based on FEV1 % Predicted

PFT results were also collected one year prior the start of the program and one year after completion of the program.

Paired t-tests were used to analyze the pre and post differences to examine the changes following completion of the program. ANCOVA tests were additionally used to test for covariate significance in changes of the pre/post data. The alpha level was set a priori at 0.05. A Bonferroni adjustment was made on alpha level threshold for the paired t-test to .025 (.05/2); this was to account for multiple tests performed.

Chapter 4: Results

Study Population

The 41 total CF patients that had been enrolled in the rehabilitation program since initiation were examined. The patients' ages ranged from 6 years to 56 years of age; 6 total patients were pediatric patients, defined as younger than 18 years of age. Eighteen patients were female, and 23 three were male. Thirty one total patients were able to complete the program; the reasons for 10 patients' incompletion of the program ranged from hospital admission, quick success in the program that made continuing unnecessary, dropping out of the program, death, or failure to participate in an exit evaluation. Days in the program ranged from 21 to 363 days, with the mean length being 63.06 days. Twelve patients were lung transplant patients; this includes both pre and post transplantation. Twenty four patients had a diabetes diagnosis. Two patients were re-enrolled into pulmonary rehabilitation and both were added to the database. Currently, 10 of the patients that underwent the pulmonary rehabilitation program are now deceased with death occurring from 1 to 36 months after rehabilitation.

Results of Research Questions

1. Does a comprehensive pulmonary rehabilitation program for adult and pediatric patients with cystic fibrosis change outcomes including:
    a. health-related quality of life

Though HRQOL data was collected, only 6 patients actually completed both pre and post survey data. This was not a large enough portion of the study population to warrant a t-test analysis. However, the descriptive data did reveal a change in the mean pre HRQOL scores to the mean post HRQOL score of 37.10 to 66.65, for the 6 total patients that completed both pre and post surveys. A higher score indicates a higher of quality of life.

The scores from a Shortness of Breath Questionnaire (SOBQ) were also collected. However, too few patients completed both the pre and post questionnaire for apparent paired t-test. The descriptive pre post data did reveal a change in the mean pre SOBQ score to the post SOBQ score of 31.125 to 20.000, for the 16 total patients that completed both pre and post surveys. A lower score indicates less shortness of breath.

The number of hospital admissions 6 months prior and post program was collected for every patient possible. Only 23 patients had both pre and post program records of hospitalizations within 6 months. The mean hospitalizations from pre-program to post program was 2.0 and 1.9 respectively, for the 23 patient that had both pre and post data recorded.

b. 6-minute walk distance

33 patients completed both a pre and post 6MW test. A paired t-test analyzed significant change in the distance walked. There was found to be a statistically significant difference in the pre/post change of the 6MW test with

p = 0.001. This shows a significant increase in distance walked in the 6MW from pre to post program. The following is a table and figure demonstrating the pre/post change.

	Pre		Post		
	Mean	Standard Error	Mean	Standard Error	p-value
6MW	**1379.5**	**63.324**	**1593.0**	**58.506**	**0.001**

*alpha threshold adjusted to .025 (.05/2) for multiple tests per Bonferroni correction

Table 2: Paired t-test for Pre/Post 6MW

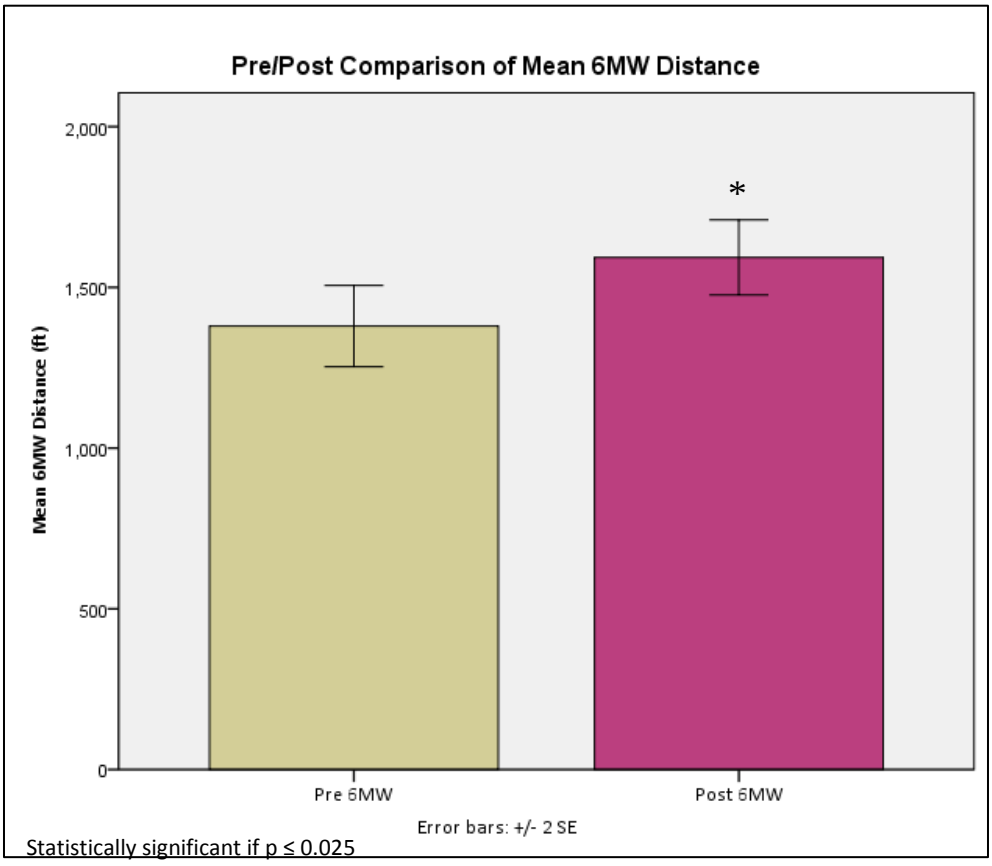

Figure 1: Pre/Post Comparison of Mean 6MW Distance

c. BMI

Data was collected to calculate a pre and post program BMI value in kg/m^2 for 36 total patients. A paired t-test was used to evaluate change. There was found to be a statistically significant difference in the pre/post change of the patients' BMI with a p = 0.002. This shows a significant gain the patients' BMI from pre to post program. The following is a table and figure demonstrating the pre post change.

	Pre		Post		
	Mean	Standard Error	Mean	Standard Error	p-value
BMI	20.	0.795	21.1	0.766	0.002

*Alpha threshold adjusted to .025 (.05/2) for multiple tests per Bonferroni correction

Table 3: Paired t-test for Pre/Post BMI

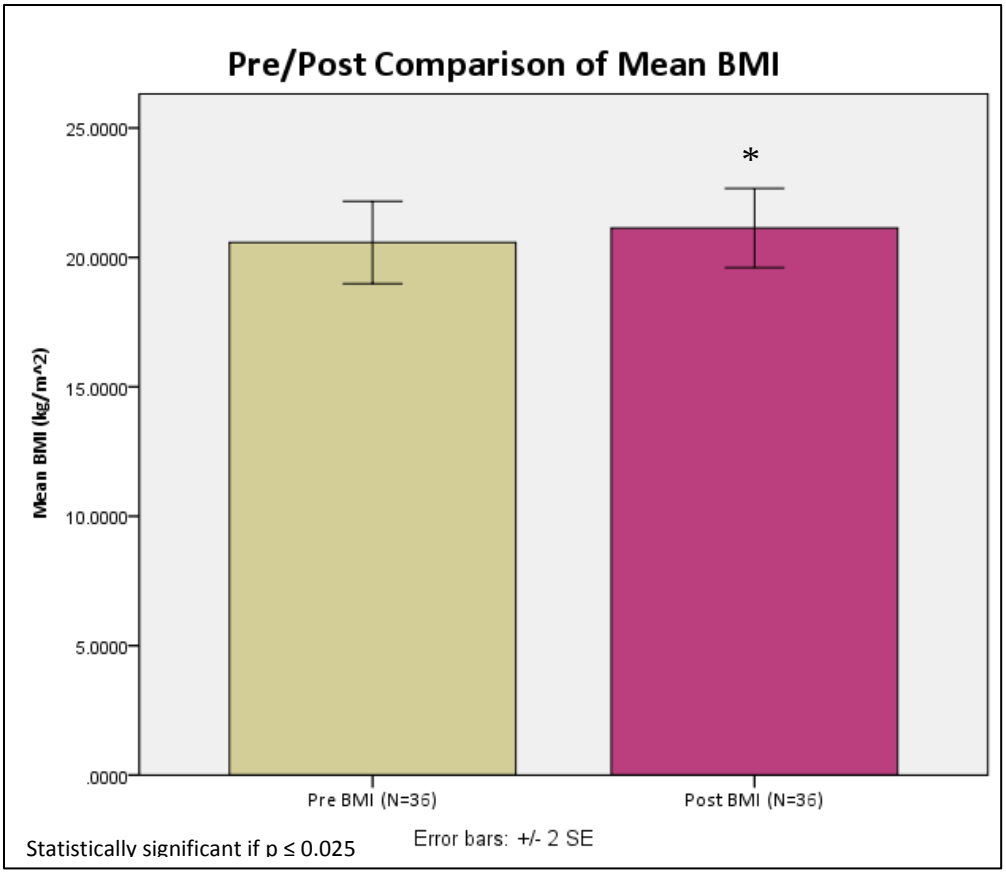

Figure 2: Pre/Post Comparison of Mean BMI

d. pulmonary function

To determine if there was an effect of the pulmonary rehabilitation program on lung function a pre/post t-test performed, the FEV1 and the FEV1/FVC to test for a significant change. Transplant patients were excluded from this analysis to control for extraneous variables, such as a lung transplant and resulting side effects or outcomes, which could also affect lung function. There was not found to be a statistically significant pre/post change in either

FEV1 or the FEV1/FVC. For the FEV1 pre/post t-test p = 0.549; p = 0.786 for the FEV1/FVC for the t-test.

	Pre		Post		
	Mean	Standard Error	Mean	Standard Error	p-value
FEV1	1.690	0.181	1.600	0.182	0.549
FEV1/FVC	0.630	0.029	0.640	0.044	0.786

*alpha threshold adjusted to .025 (.05/2) for multiple tests per Bonferroni correction

Table 4: Paired t-test for Pre/Post FEV1 and FEV1/FVC

2. Do outcomes for CF patients differ based on:

   a. age

      Due to a disproportionate number of adult patients compared to pediatric patients that completed the program, ANCOVA tests were not performed to determine if age had effect on change from pulmonary rehabilitation.

   b. disease severity

      An ANCOVA test was performed to determine if there was a significant difference between levels of disease severity on the pre/post change of the 6MW distance using an alpha threshold of 0.05. The pre/post interaction with the severity of disease was statistically significant ($F(5,27) = 2.838$, $p = 0.035$). There is a greater improvement in 6MW distance in the mild, moderate, and moderately severe levels of severity. However, this result should be interpreted with caution due to the uneven disbursement of patients across the varying levels of severity; only two patients were mild, and one

patient was moderate. The following figure demonstrates the pre/post changes in 6MW across the different levels of severity of disease.

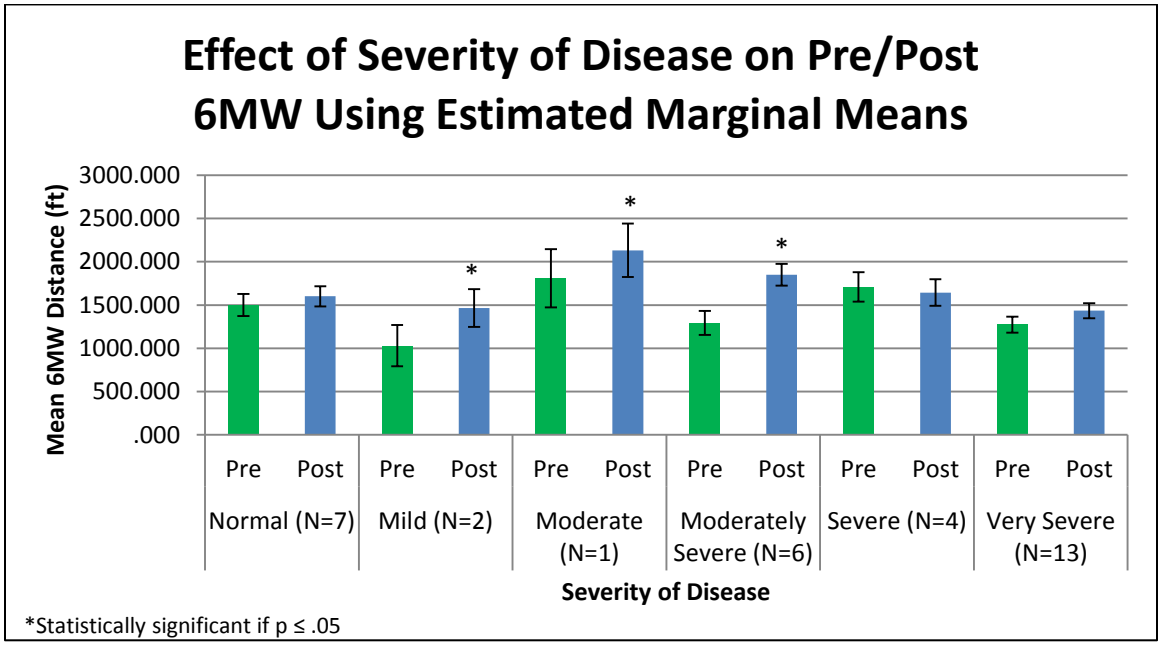

Figure 3: Effect of Severity of Disease on Pre/Post 6MW Using Estimated Marginal Means

c. program compliance

Due to incomplete records, ANCOVA tests were not performed to determine if compliance had an effect on the pre post program data.

d. sex

An ANCOVA was performed to determine if there was a significant difference between male and females on the change of BMI using an alpha ≤ 0.05. The pre/post interaction with sex was statistically significant (F (1, 34)

= 4.160, p = 0.049). There is a greater gain in BMI between the pre and post data in females than in males.

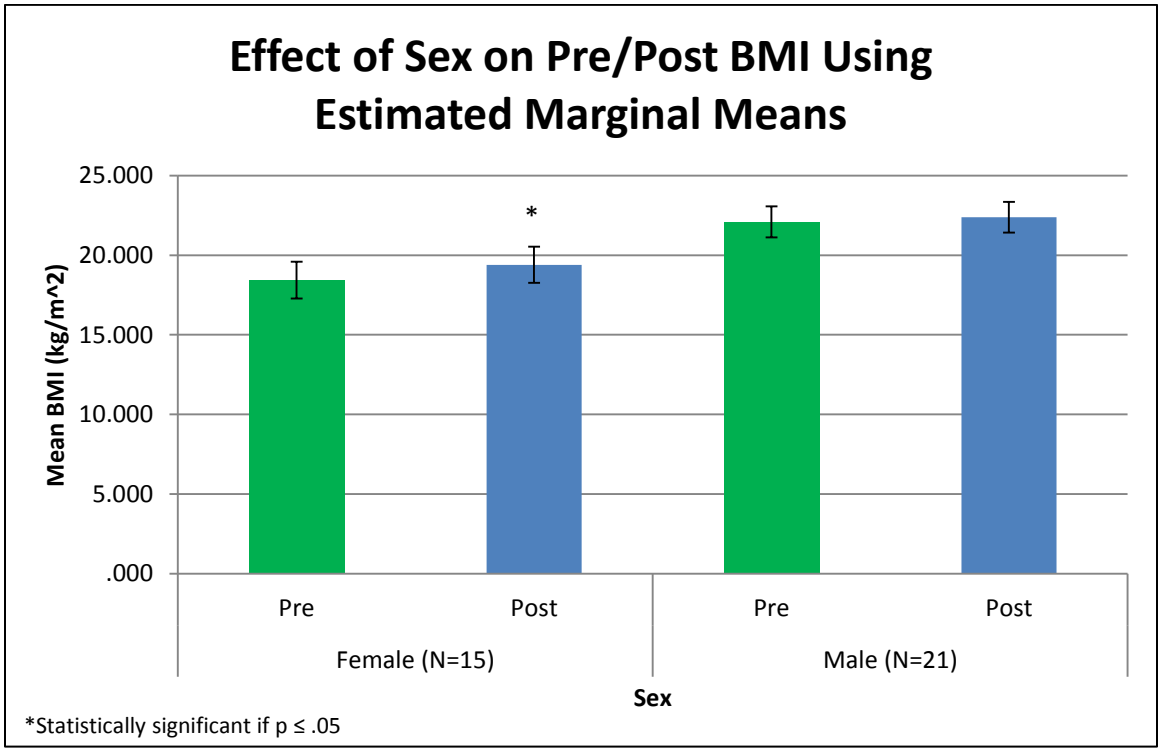

Figure 4: Effect of Sex on Pre/Post BMI Using Estimated Marginal Means

Chapter 5: Discussion

Exercise Capacity and Endurance

The significant change between the pre and post 6MW data for CF patients that engaged in the pulmonary rehabilitation program at Nationwide Children's is an indication of an increase in exercise capacity and endurance. Patients were able to walk a significantly greater distance after completion of the program compared to at the start in the same six minute interval of time. It has been previously illustrated by Nixon in 1992 that aerobic fitness is strongly correlated with 8 year survival rates in CF patients and though survival rates of the patients in the program have not been closely analyzed, exercise through pulmonary rehabilitation has been shown to be beneficial to this patient population evident by the significant change in walk distance. This increase in exercise capacity and endurance in CF patients due to pulmonary rehabilitation is consistent with results from research studies dating back to the mid-1970s that have shown that pulmonary rehabilitation programs have had similar successes in increasing exercise tolerance in COPD patients (Troosters et al, 2005). Increased exercise capacity could also lead to decreased dyspnea. Compiled descriptive data of the scores from the pre and post SOBQ did show a positive change in reduced shortness of breath after completion of the program for the patients that did complete this survey. This reduction of dyspnea is consistent with another shown benefit of pulmonary rehabilitation in the COPD population by the California Pulmonary Rehabilitation Collaborative Group in 2004.

Lung Function

This study also examined if pulmonary rehabilitation caused some significant change in PFT results which are evidence of lung impairment and function. This portion of the study excluded patients that received lung transplants to control for extraneous variables due to transplantation. Because there was no significant change in FEV1 or FEV1/FVC values found between the pre and post data, this is an indication that lung impairment by obstruction was unaffected by pulmonary rehabilitation. This supports research found Moeller et al. in 2010 that demonstrated in a short term inpatient rehabilitation program there was no significant effect of pulmonary rehabilitation on overall lung obstruction. The pulmonary rehabilitation was also relatively short in length for most patients with a mean length of 63.06 days. However, Moller et al. also found in 2010 a significant change of FVC and IC in pre post data from a short term inpatient rehabilitation program. Unfortunately IC data could not be collected in this study as Nationwide Children's does not generally obtain an IC value. Descriptive data was compiled for the FVC excluding transplant patients, and showed a decrease from pre-program (2.6541) to post program (2.4841); paired t-tests were not performed. This decrease is not consistent with previously published literature. This study does not support that pulmonary rehabilitation positively or negatively affects lung function. However, as emphasized by Mathews et al. in 1964, CF is a genetically inherited, chronic disease with no known cure; therefore, it is reasonable that improved exercised capacity does not necessarily correlate with improved lung function in this patient population. However, this study did find there to be a significant difference between patients'

improvement in exercise capacity based on the severity of their disease state determined based on lung function tests.

Patients' severity of disease was classified based on their pre FEV1 % predicted PFT values into classifications of normal, mild, moderate, moderately-severe, severe, or very severe. There was found to be a significant difference between pre and post 6MW distances for the varying levels of disease severity. Patients with mild, moderate, and moderately-severe lung impairment had greater changes in their 6MW from pre to post program than patients with normal, severe, or very severe lung impairment. Though this result should be interpreted with caution due to the uneven disbursement of patients across the varying classifications of severity; two patients had mild lung impairment, and one had moderate impairment, this demonstrates that pulmonary rehabilitation is most beneficial in patients with some deterioration of lung function, but not as beneficial in patients with normal, severe, or very severe lung impairment. Patients with normal lung function are not in need of pulmonary rehabilitation such as patients with lung impairment. Patients with severe or very severe lung function are patients that are probably in consideration for more drastic measures of treatment such as lung transplantation. The increased severity of their disease could also impair patients' engagement in the physical activities of pulmonary rehabilitation compared to patients with less lung function deterioration.

Of the 17 patients in this study that fell into both the severe and very severe lung impairment categories, five of those patients died within 1 to 36 months, additionally, 3 patients received lung transplantation. Bradley, J., & Moran, F. published in 2008 that

physical exercise may slow the deterioration in lung function in CF patients. Based on results from this study, if physical exercise does slow deterioration in lung function it is not as likely in patients currently suffering from severe or very severe lung impairment.

BMI and Weight Gain

A significant change in BMI from the pre to post data indicates that the program was successful in increasing patients' weight, which is usually a goal for CF patients due to the general difficulties in gaining for weight for this patient population. BMI was analyzed instead of weight because this study included both pediatric and adult patients; therefore height was a controlled variable, which is especially important in growing pediatric patients. An increase in BMI could be attributed to various elements of the pulmonary rehabilitation program, such as physical therapy, therapeutic recreation/occupational therapy, nutrition, or education. It is likely due to the multidisciplinary approach that helps these patients increase weight gain, by developing muscle mass, learning the proper foods to consume, and becoming educated in breathing techniques to conserve energy. This statistically significant increase in BMI from pre to post program is supported by previous research published by Orenstein et al. in 2004 who found CF patients that engaged in either aerobic or anaerobic exercise had a significant increase in weight after 6 and 12 months of exercising. This study found that females with CF increased their BMI pre to post program significantly more than males with CF that enrolled in the program. This result shouldn't discourage CF pulmonary rehabilitation programs from enrolling male patients; the male CF patients did show an

increase in BMI pre to post program. Additionally there are other benefits of pulmonary rehabilitation for CF patients outside of weight gain.

Heath Related Quality of Life

A primary goal of pulmonary rehabilitation is to improve patients' functional and health related quality of life. Like any patient with a chronic disease state, CF patients face hardships from their physical limitations which can lead to a deterioration of quality of life. Nationwide Children's pulmonary rehabilitation program aims to improve patients health related quality of life through physical training, to help patients physically cope with disease process, and through consultations with psychology to help patients mentally cope with symptoms and to generally improve their emotional mental status. There were insufficient pre and post HRQOL surveys collected from patients that completed the program available to run paired t-tests. However, there was a difference between the pre (37.10) and post (66.65) mean survey score for the 6 patients that did complete both the pre and post surveys. This promising increase in scores indicates that patients had an increase in health related quality of life after completion of the program. The improvement in quality of life in CF pulmonary rehabilitation patients is also supported in previous literature regarding improved quality of life in COPD pulmonary rehabilitation patients (Troosters et al, 2005). Consistent with Griese's et al. in 2010, struggle with poor record keeping, this study also supports the need for consistent and improved record keeping. Improving health related quality of life is an important element of pulmonary rehabilitation that should be reexamined at Nationwide Children's after a few years of improved documentation and record keeping.

Reduction of Hospital Admissions

As healthcare professionals, the government, and hospitals work to drive down healthcare costs, there has been a recent push to reduced readmission rates. A primary goal of all pulmonary rehabilitation programs based on their definition has been to "reduce health care costs through stabilizing or reversing systemic manifestations of the disease" (Nici et al, 2006). Therefore, in addition to the data collected to answer research question, the number of hospital admissions 6 months prior and post program was investigated for every patient. The hospitalizations recorded were limited to admissions in Nationwide Children's; admissions to outside hospitals were not recorded. Additionally, there was not data available to collect the number of hospitalizations for every patient; only 23 patient had both pre and post hospitalization data recorded. There was only a slight decrease in mean hospitalizations from 2.00 pre-program, to 1.91 post program. The change was slight so paired t-tests were not performed. Hospitalizations and admissions are important data to collect by pulmonary rehabilitation programs. Even a slight decrease in the number of hospitalization post pulmonary rehabilitation could be substantial in improved patient quality of life and reduced health care cost from hospital admission; this could be a great benefit of pulmonary rehabilitation with CF patients.

Conclusion

Based on this study, it is recommended that Nationwide Children's and all other pulmonary rehabilitation programs should keep consistent and complete records for all patients. Based on the improved outcome measures found, this study demonstrates the benefits of pulmonary rehabilitation in the CF population. Implications for clinical

practice could include the creation of more pulmonary rehabilitation programs aimed towards CF patients, increased referrals for CF patients to pulmonary rehabilitation programs, and increased insurance reimbursement for CF patients enrolled in pulmonary rehabilitation programs. Ultimately, this study supports and recommends the use of pulmonary rehabilitation with CF patients.

Recommendations for Future Research

Future studies should examine the HRQOL in CF patients enrolled in pulmonary rehabilitation, as well as examine the number hospitalizations of CF patient's pre and post pulmonary rehabilitation. Additional studies could also monitor the effects more closely of pulmonary rehabilitation in the recovery of lung transplant in CF patients.

Limitations

Limitations to this study were incomplete records and inconsistencies in the program between patients, such data collected, duration of the program, and surveys completed. An additional limitation was the population size, especially in the pediatric patient population.

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